Specialty Update

What's New in Limb Lengthening and Deformity Correction

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This update summarizes select articles pertaining to limb lengthening and deformity correction that were published between July 1, 2011 and December 31, 2012.

Limb-Length Discrepancy

The incidence of abdominal neoplasms was 10% (one of ten) in children with syndromic hemihypertrophy and 1.2% (three of 250) in those with isolated idiopathic hemihypertrophy[1]. Limb-length discrepancy can adversely affect the spine and pelvis[2]. On the basis of a finite element study, Kiapour et al. found that an incremental increase in limb-length discrepancy was associated with increasing stress across the sacroiliac joint[3]. Laser-based ultrasound was an accurate and reliable noninvasive modality for assessing limb-length discrepancy in young adults[4]. In a study examining parental concerns regarding their child's limb-length discrepancy, Lee at al. noted that a discrepancy of >2 cm was associated with the willingness of parents to seek treatment for their child[5].

Pediatric Lower Extremity Disorders

A study comparing the limb lengthening achieved with use of monolateral or circular fixators found similar healing indices[6]. While the Taylor spatial frame (Smith & Nephew, Memphis, Tennessee) was most accurate in achieving deformity correction and was associated with a lower complication rate, it was a more expensive device[6]. The technique of circumferentially dividing and stripping the periosteum of the short femoral and tibial diaphyses for addressing <6 cm of limb-length discrepancy in young children appears promising[7].

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The distal fibular physis is typically unaffected in meningococcal septicemia, creating varus angulation at the ankle[8]. Among children with massive bone defects secondary to chronic osteomyelitis of the tibia, distraction osteogenesis by means of external fixation is an effective technique[9]. Distal tibial valgus can be addressed with use of plate hemiepiphysiodesis as an alternative to the transmalleolar screw technique[10]. Percutaneous epiphysiodesis with use of transphyseal screws at the proximal part of the tibia was associated with a substantial rate of undercorrection and need for revision of the hardware[11]. Standard 3.5-mm stainless steel plates fixed with noncannulated screws can be used as an alternative to specific guided growth implants for correcting angular deformities about the knee[12,13].

On the basis of magnetic resonance imaging and comparison with control patients, patients with Blount disease were found to have a thicker proximal medial tibial chondroepiphysis, a thicker and wider medial meniscus, and greater frequency of abnormal signals in the posterior horn of the medial meniscus[14]. Use of guided growth for children with infantile Blount disease was effective in correcting varus malalignment but was also associated with several cases of screw breakage (17%) and wound infection (17%)[15].

Arthroscopic evaluation of the knee in twenty-one patients with proximal femoral focal deficiency revealed absence or hypoplasia of one or both cruciate ligaments in 95% of patients[16]. Despite such findings, patients with fibular hemimelia and anterior cruciate ligament deficiency can maintain active lifestyles, similar to age-matched controls[17]. Varus overcorrection was recommended when addressing genu valgum in patients with congenital lower limb deficiency and ball-and-socket ankle joints[18].

Gradual distraction with use of circular external fixation is an effective treatment for relapsed or neglected clubfeet[19-22],

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including those with arthrogryposis[23]. Guided growth modulation with use of anterior distal tibial epiphysiodesis did not improve ankle dorsiflexion among children with clubfeet who had a recurrent equinus deformity[24].

For patients with atrophic congenital pseudarthrosis of the tibia and fibula, creation of a large cross-sectional area of osseous union with primary osteosynthesis of the tibia and fibula was recommended[25]. Agashe et al. reported ultimate union in fourteen of fifteen children with atrophic congenital pseudarthrosis of the tibia who underwent the traditional Ilizarov technique combined with antegrade intramedullary nailing[26]. Preliminary data on sixteen children undergoing free fibular transfer for atrophic congenital pseudarthrosis of the tibia demonstrated primary healing in 37% of the patients, with a 37% refracture rate and 37% rate of recurrence of the apex anterior deformity[27]. When a free fibular transfer is performed, at least 5 cm of distal fibular remnant should be retained in the donor leg to prevent an iatrogenic valgus deformity of the ankle[28].

For patients with late-onset (i.e., onset after eight years of age) Legg-Calvé-Perthes disease, hip distraction with use of external fixation led to improvement in joint mobility and decreased pain[29-31]. A two-stage management protocol with initial iliofemoral distraction followed by open reduction and pelvic osteotomy demonstrated promising results for eight to ten-year-old children with previously untreated unilateral high developmental hip dislocations[32].

In patients with multiple epiphyseal dysplasia, radiographic measurement of the tibiofemoral angle and mechanical axis deviation is reliable[33]. For children with chondrodysplasia and hip and knee varus, improved alignment can be achieved with multilevel guided growth treatment at the lateral portion of the proximal part of the femur, distal part of the femur, and proximal tibial physes[34]. Recurrence following correction of genu valgum was noted in eight of twenty children with Morquio syndrome[35]. Equinus contracture and recurrent genu varum are often noted in patients with skeletal dysplasia who are undergoing gradual correction with use of the Ilizarov method[36]. In twenty-two patients with osteogenesis imperfecta who were followed for an average of nineteen years following insertion of Sheffield telescopic intramedullary rods, lack of physeal damage and anticipated elongation of the implant with growth were documented at a nineteen-year mean follow-up. However, a 50% reoperation rate related to implant-related issues was noted[37].

Upper Extremity Reconstruction

Gradual correction with external fixation can be used to address shortening and angular deformities of the upper extremity. Pawar et al. noted that humeral lengthening with use of monolateral external fixators was well tolerated and effective in improving function, with complication rates similar to those reported with circular frames[38]. The use of multiplanar external fixators with computer-based programming for upper extremity reconstruction is also gaining popularity[39]. Hill et al. reported on the use of distraction osteogenesis to restore radioulnar length or to lengthen both bones of the forearm in nineteen children with various diagnoses[40]. While such techniques may improve function and cosmesis, pin-site infections and poor regenerate formation are common[40]. The need for ulnar lengthening in children with forearm deformities secondary to multiple hereditary exostoses remains controversial[40-42]. Litzelmann et al. reported reasonable function at skeletal maturity despite major radiographic abnormalities and limited functional gain following ulnar lengthening[41]. While lengthening of the shortened ulna is an established procedure for children with congenital radial longitudinal deficiency, reports of untoward events such as recurrent deformity[43] and growth retardation of the lengthened ulna[44] persist. Use of elastic intramedullary nailing to supplement gradual lengthening of the forearm can shorten the external fixation time with minimal additional morbidity[45]. Forearm shortening osteotomy and volar radiocarpal capsulotomy to address flexion deformities of the wrist in children with amyoplasia led to recurrent deformity with no substantial gain in function[46].

Trauma

The ability to rapidly stabilize injuries of the extremities with minimal additional soft-tissue trauma has made external fixation an integral part of damage-control orthopaedics[47]. Gordon et al.[48] reviewed the principles of applying external fixators in combat and austere settings. The use of safe corridors during insertion of half-pins can minimize iatrogenic injury. In a cadaveric study, Beltran et al.[49] noted the vulnerability to muscular branches of the femoral nerve and the suprapatellar pouch during insertion of half-pins in the anterior aspect of the femur. If clinically feasible, anterolateral or lateral pin placement was recommended. In a report on battlefield injuries, early conversion from external to internal fixation provided rapid healing without infection, whereas cases of late conversion were associated with longer time to union and increased deep infections[50].

Application of a programmable circular fixator in ten children with high-energy open tibial shaft fractures allowed early weight-bearing and satisfactory clinical outcome[51]. The use of circular external fixators to treat forty adults who had complex acute tibial shaft fractures (including nineteen open fractures) led to primary healing and satisfactory outcome in the majority of patients[52]. Applying articulated circular fixation in patients with complex knee dislocations can facilitate early mobilization following multiligament repair and/or reconstruction[53]. The use of a novel multiplanar external fixator with percutaneous Kirschner wires following indirect closed reduction of complex proximal humeral fractures allowed early mobilization while minimizing the risk of osteonecrosis and deep infection[54].

Compared with a midline triceps-splitting approach, a lateral, triceps-sparing approach was associated with a lower rate of postoperative neurologic deficits[55] as well as a lower rate

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of loss of elbow mobility[56] following a supracondylar osteotomy for malunited fractures in children. Use of three-dimensional computed tomography scanning may facilitate preoperative planning in such cases[57]. Corrective osteotomy for posttraumatic cubitus varus in children with use of a locking lateral closing-wedge osteotomy was described by Skaggs et al.[58]. In adults, more stable fixation with use of dual plating is recommended for these osteotomies[59].

Use of a computer-assisted system, including a custom-made osteotomy template, can aid in the surgical correction of malunited diaphyseal fractures of the forearm[60]. The ideal technique for correction of pediatric distal radial malunions secondary to premature physeal closure remains controversial. Both acute lengthening with tricortical graft and plate fixation[61] and external fixation with use of distraction osteogenesis[62] have been successful. For adult patients with distal radial malunions, a closing-wedge osteotomy with concomitant distal ulnar shortening yielded better restoration of alignment, wrist mobility, and function than did an opening-wedge correction[63]. The use of supplemental allograft when performing distal radial osteotomies with plate fixation did not improve clinical outcome[64].

Bone transport with use of external fixation continues to be effective as a treatment method for adult traumatic long-bone defects and as a salvage method in patients who have undergone removal of a megaprosthesis as a result of infection[65-68]. Among children, sizable bone defects with a preserved periosteal sleeve can heal after application of massive amounts of autograft bone in the skeletal gap[69]. In biologically unfavorable scenarios, atrophic tibial nonunions in pediatric patients can be successfully treated with a vascularized periosteal flap from the fibula[70]. Bumbaširević et al. used a percutaneous distraction-compression technique with a fine-wire Ilizarov fixator to treat scaphoid nonunion in eighteen adult patients without associated deformity or osteonecrosis[71]. The Ilizarov method has also been used to treat septic nonunion of the distal part of the humerus[72].

Lower Extremity Reconstruction

Malalignment of the tibial and femoral components during total knee arthroplasty, especially in patients with limb malalignment and morbid obesity, can affect the contact forces at the implant-bone interface and lead to early implant failure[73,74]. In patients with genu varum who undergo total knee arthroplasty, varus alignment of the tibial component can negatively impact clinical outcome and increase the possibility of requiring revision arthroplasty[75]. Mullaji et al. noted that, in patients undergoing total knee arthroplasty, excessive preoperative malalignment was associated with postoperative malalignment[76].

The authors of a Finnish registry-based study found that survivorship following high tibial osteotomy for osteoarthritis was 89% at five years and 73% at ten years[77]. The authors of another large study noted a 70% survival rate at ten years following high tibial osteotomy[78]. The performance of an opening-wedge high tibial osteotomy with use of hemicallotasis via external fixation for the treatment of medial compartment arthritis can provide precise correction without altering the tibial slope or patellar height[79]. Infusion of zoledronic acid during the consolidation phase for patients undergoing high tibial osteotomy via hemicallotasis did not enhance healing time[80]. The outcome of subsequent total knee replacement was not adversely affected by a previous high tibial osteotomy[81]. Distraction arthroplasty (arthrodiastasis) may be a useful adjuvant to arthroscopic lavage and drilling in osteoarthritis[82]. However, further investigation is warranted to better define the role of arthrodiastasis in patients with osteoarthritis of the knee.

Pelvic support osteotomy with ipsilateral distal femoral lengthening (Ilizarov hip reconstruction) was used to address pain and gait deviations in patients with instability of the hip joint related to various etiologies[83-85] and in patients with advanced osteonecrosis following leukemia[86].

Foot and Ankle

For certain patients with painful arthritis, the use of controlled ankle-joint distraction via external fixation is gaining popularity[87,88]. In a randomized controlled trial, Saltzman et al. noted that, in comparison with fixed-joint distraction, application of a hinged distraction frame provided a better clinical outcome in patients with symptomatic osteoarthritis of the ankle[89].

Malunited ankle fractures and asymmetric arthritis of the ankle can be corrected with use of a supramalleolar osteotomy, with good-to-excellent outcome in the majority of patients[90,91]. The use of the Taylor spatial frame is an effective technique to gradually correct multiplanar deformities of the distal part of the tibia[92,93]. Lee et al. noted that radiographic alignment of the tibial plafond varied greatly among patients with early stage arthritis in the medial aspect of the ankle[94]. Barg et al. questioned the reliability of assessing distal tibial alignment with use of weight-bearing radiographs of the ankle and hindfoot[95]. A cadaveric study demonstrated that, with an intact fibula, a supramalleolar osteotomy producing varus alignment overloaded the lateral aspect of the tibiotalar joint, while a valgus angulation overloaded the medial aspect of the joint. These biomechanical effects were reversed after the performance of a distal fibular osteotomy proximal to the syndesmosis[96]. An intra-articular opening-wedge osteotomy of the tibial plafond and lateral ligament reconstruction can be used to address unstable ankle joints with intra-articular varus deformity and arthritis[97].

A plantigrade position of the foot was achieved with use of a U-osteotomy with distraction osteogenesis in fifteen patients with a stiff foot with multiplanar deformities[98]. A V-shaped double osteotomy was useful for simultaneous corrections of select midfoot and hindfoot deformities[99]. A staged technique with initial external fixation to achieve equinocavovarus deformity correction followed by percutaneous screw fixation and frame removal was well tolerated[100].

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Use of a circular fixator to achieve ankle arthrodesis is effective in managing patients with end-stage osteoarthritis associated with severe deformity, bone loss, or infection[101-103]. In a large series, smokers and those with Charcot arthropathy had worse clinical outcomes[102]. In another study, a fixator-assisted arthrodesis was effective in managing individuals with failed ankle arthroplasty and secondary bone loss[103]. In patients with severe valgus deformities who were candidates for hindfoot arthrodesis, a medial surgical approach was safe and effective[104].

Oncologic Reconstruction

In addition to allografts and various endoprosthetic devices, limb reconstruction with use of distraction osteogenesis can be a viable biologic salvage option for managing malignant and aggressive benign bone tumors. Cañadell's technique of physeal distraction facilitated the resection of malignant metaphyseal bone tumors, despite their close proximity to an open physis[105]. Following oncologic resection, bone transport or acute shortening of the surgical defect with gradual lengthening at a distant portion of the same bone was used to reconstruct skeletal defects in patients with intact epiphyses. Despite prolonged external fixation time, satisfactory functional outcome with this technique was reported in the majority of patients from two different centers[106,107]. Another innovative method of managing intercalary skeletal defects following oncologic resection involved a two-stage technique in which a temporary methyl methacrylate cement spacer was used to produce a bioactive membrane, followed four to eight weeks later with placement of cancellous autograft within this membrane[108,109].

Stature Lengthening

Bilateral lower limb lengthening for increasing standing height remains controversial. A recent report suggested that the quality of life of patients with achondroplasia improved following stature lengthening[110]. However, with extensive lengthening in such children, growth inhibition of several centimeters has been documented[111,112]. Among patients with achondroplasia, humeral lengthening (as opposed to femoral lengthening) was associated with a lower complication rate, faster consolidation, and better quality of some quality-of-life metrics[113]. Other authors have reported using lengthening over an intramedullary nail[114] and lengthening and then intramedullary nailing[115] for patients undergoing stature lengthening.

Postoperative Complications and Their Management

The field of limb lengthening and deformity correction is associated with several pitfalls and complications. However, with careful patient selection, preoperative planning, and postoperative vigilance, these untoward events can be minimized. Koyonos et al. recently reviewed the potential errors and the strategies for prevention and management of complications pertaining to osteotomies about the knee[116]. In a study comparing epidural analgesia, intravenous morphine infusion, and continuous peripheral nerve blocks postoperatively in patients undergoing limb reconstruction with circular frames, sciatic nerve catheterization provided the most effective analgesia[117]. However, close vigilance is necessary to avoid missing a compartment syndrome postoperatively.

Neurovascular injury as well as contractures and instability of adjacent joints during distraction osteogenesis can lead to devastating consequences. The intraoperative use of somatosensory evoked potentials during external fixation demonstrated 100% sensitivity and 91% specificity for the detection of nerve injury in children undergoing limb lengthening and deformity correction[118]. Equinus contracture developed in eight (7.8%) of 102 patients undergoing tibial lengthening, despite the use of an ankle-foot orthosis for at least sixteen hours per day[119]. Less than 10° of sagittal plane deformity was often noted following a femoral osteotomy and did not substantially increase with progressive lengthening[120]. In another retrospective case series, hip dislocation during extensive lengthening of a congenitally short femur was associated with progressive acetabular dysplasia. Closed reduction was ineffective in three such children, and soft-tissue releases and open reduction with femoral shortening plus acetabular reconstruction were ultimately necessary for joint relocation[121].

Despite advances in technology, pin-track infection remains a relatively common problem in patients with external fixators. Various strategies have been proposed for preventing and treating such pin-related issues, including ensuring stability at the pin-bone interface through the use of hydroxyapatite-coated half-pins, avoiding thermal necrosis during pin insertion, using various types of dressings around the pin sites, and liberal use of oral antibiotics to manage early pin-track infections[122-127]. On the basis of a randomized controlled trial, Lee et al. noted that, compared with the use of saline solution, the use of polyhexamethylene biguanide-impregnated gauze reduced the risk of pin-track infections in patients undergoing limb lengthening and deformity correction[126].

Methods to enhance new bone formation during distraction osteogenesis continue to be studied. Latalski et al. found that platelet-rich plasma injected into the regenerate bone at the end of the distraction phase was effective in decreasing the duration of external fixation in patients undergoing limb lengthening[128]. On the basis of a randomized unblinded controlled trial, Dudda et al. noted that, although the application of low-intensity pulsed ultrasound did not produce a significant decrease in the distraction-consolidation index (32.8 days of treatment per centimeter of lengthening in the study group as compared with 44.6 days of treatment per centimeter of lengthening in the control group, $p = 0.116$), the fixator gestation period could be decreased by approximately forty-four days in the treatment group[129].

It is important to maintain stability of the external fixator, including during strut changes, when the Taylor spatial

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frame is being used. Jenkins at al recommended placing the temporary "seventh" strut in the same orientation as the strut that is being exchanged[130].

New Tools and Techniques

Digital radiography and low-dose imaging are reliable modalities for the assessment of mechanical axis deviation of the lower extremity[131,132]. A spirit-level mounted on circular fixators can be used to accurately position the limb for postoperative radiography[133]. New bone formation during distraction osteogenesis can be quantified by assessing bone mineral density with use of dual x-ray absorptiometry scanning and the measurement of pixel value ratios of digital radiographs[134,135].

Designs of intramedullary lengthening devices continue to evolve. While these implants may not have some of the problems associated with external fixators[136], other unique device-related issues continue to surface. In patients undergoing femoral lengthening with use of intramedullary telescopic nails, approximately 1 mm of lateral shift of the mechanical axis occurs for each 1 cm of lengthening due to lengthening along the anatomical axis (rather than mechanical axis) of the femur[137]. The authors of a case series of fourteen patients undergoing lower limb lengthening with use of a motorized intramedullary nail reported complications that included malfunction of the distraction mechanism, delayed consolidation, valgus deformity, and femoral fractures at the proximal tip of the nail[138]. In another report of thirty-six patients undergoing femoral lengthening with use of a telescopic nail, although bone consolidation was achieved faster with the telescopic nail than with external fixation, 17% of patients required a second procedure to achieve the lengthening and 22% required one or more unplanned device manipulations with the patient under general anesthesia[139]. Among sixteen patients who underwent lower limb lengthening with another commercially available intramedullary lengthening device, 38% had poor regenerate requiring bone-grafting and 19% had nails that were difficult to distract, necessitating closed manipulation with the patient under anesthesia[140]. Thus, it is prudent to counsel patients regarding the possibilities of encountering major obstacles, such as device failure and multiple returns to the operating room, when such intramedullary lengthening devices are used[141].

A technique for addressing femoral fracture malunion and resultant limb-length discrepancy through the application of principles of lengthening over an existing intramedullary nail was reported[142]. In another report, in comparison with use of an intramedullary lengthening nail, the use of femoral lengthening over a nail was associated with fewer complications and a more controlled distraction rate[143]. However, there is a higher risk of the development of deep infection with use of the technique of lengthening over a nail[144]. In addition to being associated with infection, tibial lengthening over a nail can be associated with valgus angulations of the tibia (25%) and equinus contracture (72%)[145].

A minimally invasive technique for acutely correcting distal femoral valgus deformities with fixator-assisted locked plating was described[146]. Accurate correction of the distal femoral deformities was obtained with both fixator-assisted plating and conventional external fixation techniques[147]. The external fixation time associated with lengthening and then plating was shorter than the time associated with conventional lengthening with external fixation, but varus deformity and implant failure were observed in the group of patients who underwent lengthening and then plating[148].

Upcoming Events

Specialty Day of the Limb Lengthening and Reconstruction Society (LLRS) will be held at the Annual Meeting of the American Academy of Orthopaedic Surgeons (AAOS) on March 15, 2014, in New Orleans, Louisiana. The Annual Scientific Meeting of the LLRS will be held on July 25 and 26, 2014, in Montreal, Quebec, Canada. Details are available at the LLRS web site: www.llrs.org.

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